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## Doença falciforme: perspectivas sobre assistência prestada na atenção primária\*

Sickle cell disease: perspectives on the assistance provided in primary attention

Anemia de células falciformes: perspectivas sobre assistência prestada en la atención primaria

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### ABSTRACT

**Objective:** To investigate the vision of patients with sickle cell disease on assistance provided in the Family Health Units in accordance with the decree 1,391/05. **Methods:** Cross-sectional, descriptive, quanti-qualitative study; conducted with twenty six patients with sickle cell disease treated at Hemope. The study was approved by the CEP with CAAE: 45243015.7.0000.5195. **Results:** There was a dissatisfaction among people with sickle cell disease towards the care that is provided to them in primary attention, leading to a low accreditation that the service has on the population. **Conclusion:** The premises established by Ordinance GM 1391/05 have not been met after ten years of its implementation. Public policies aimed at black people need to be rescued so that there is continuity of care.

**Descriptors:** Sickle Cell Anemia; Primary Health Care; Public Health; Family Health.

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## RESUMO

**Objetivo:** Investigar a visão dos portadores de doença falciforme sobre a assistência prestada nas Unidades de Saúde da Família de acordo com a Portaria 1.391/05. **Métodos:** Estudo transversal, descritivo, quantitativo; realizado com vinte e seis portadores de doença falciforme atendidos no Hemope. O estudo foi aprovado pelo CEP com CAAE: 45243015.7.0000.5195. **Resultados:** Verificou-se insatisfação por parte dos portadores de doença falciforme para com o atendimento que lhes é prestado na atenção primária, levando a uma baixa acreditação que o serviço possui diante da população. **Conclusão:** As premissas estabelecidas pela Portaria GM 1.391/05 ainda não foram atingidas após dez anos de sua implantação. As políticas públicas voltadas à população negra precisam ser resgatadas para que exista uma continuidade da assistência. **Descritores:** Anemia Falciforme; Atenção Primária à Saúde; Saúde Pública; Saúde da Família.

## RESUMEN

**Objective:** Investigar la visión de los pacientes con enfermedad de células falciformes sobre la asistencia prestada en las Unidades de Salud de la Familia, de acuerdo con el Decreto 1,391/05. **Métodos:** Estudio transversal, descriptivo, cuantitativo y cualitativo; realizado con veintiséis pacientes con enfermedad de células falciformes tratados en Hemope. El estudio fue aprobado por el CEP con CAAE: 45243015.7.0000.5195. **Resultados:** Hubo descontento entre las personas con enfermedad de células falciformes hacia la atención que se proporciona a ellos en la atención primaria, lo que lleva a una acreditación bajo que el servicio tiene en la población. **Conclusión:** Las premisas establecidas por la Ordenanza GM 1391-1305 no se han cumplido después de diez años de su aplicación. Las políticas públicas dirigidas a las personas negras tienen que ser rescatados para que haya continuidad de la atención. **Descriptores:** Anemia de Células Falciformes; Atención Primaria a la Salud; Salud Pública; Salud de la Familia.

## INTRODUCTION

Sickle cell disease (DF) is one of the oldest hereditary diseases in mankind, characterized by the presence of the HbSS genotype and the formation of sickle-like red blood cells, due to the substitution of an amino acid in the hemoglobin molecule. It is most commonly found on the African continent, but with forced immigration as a result of slavery, SCD eventually spread throughout the national territory, making it one of the most frequent genetic alterations in Brazil.<sup>1,2,3</sup>

The acute symptoms of the pathology are mainly caused by the obstruction of the blood vessels by sickle-shaped red blood cells, generating hypoxia in the tissues and painful crises in the abdominal region, lungs, joints and bones. The spleen is one of the organs most affected by the obstruction of the blood vessels, causing in the loss of its function during the first years of the childhood.<sup>3</sup>

On the other hand, the chronic symptoms derive especially from the hypoxia injuries in the tissues, such as renal and cardiac insufficiency, ulcers, necrosis in bones (particularly in the humerus and femur heads) and ocular lesions.<sup>3</sup>

For the year of 2013, it was estimated the existence of more than 27 thousand Brazilians with SCD, making this pathology to be considered as a public health problem. In the state of Pernambuco, it was estimated that there were 2,000 patients with hemoglobinopathy.<sup>4</sup>

Because it is a chronic pathology, SCD therapy is based on the prevention of complications and the treatment of diseases. The look should encompass general care for monitoring growth, somatic development, psychological and organic lesions, as well as specific comorbidities.<sup>5</sup>

Thus, the importance of a multiprofessional team trained and of adequate assistance, both in basic and specialized care, is the guarantee of success in the treatment of SCD in countries such as the United States and Cuba, where public policies of attention to the sickle cell disease were implanted more than 35 years ago, resulting in increased longevity for these patients.<sup>1</sup>

Thus, considering the need for multiprofessional follow-up and aiming at a better care for patients with SCD, it was established the Administrative Rule GM 1,391/05, which guarantees comprehensive care for these patients, highlighting the concept of preventive medicine established by Leavell and Clark (1976) that is divided into five distinct components: promotion, prevention, early diagnosis, treatment and rehabilitation of health problems.<sup>6,7</sup>

The Administrative Order GM 1,391/05 recommends that the Unified Health System should promote the follow-up of people diagnosed with SCD, receive them and integrate them into the care network, as well as ensure the integrality of care, through the assistance performed by staff multidisciplinary; establish a training policy for all actors involved, promote lifelong learning, access to information and genetic counseling; ensure essential medicines; and stimulate research, with the aim of improving the quality of life of these individuals.<sup>6</sup>

However, after almost a decade of implementation of this ordinance, there are still difficulties in the care of patients with SCD in the scope of primary care, highlighting the lack of knowledge about this pathology and its treatment methods, which may lead to a discrediting of the system by the client.<sup>8</sup>

Based on the assumption that the Family Health Strategy is the preferred gateway to the Unified Health System (SUS), and that it came with a view to reorganizing the health care model with a focus on health promotion and prevention, the objective of this study was to investigate the vision of patients with sickle cell disease on the care provided at the Family Health Units (FHUs) in detriment of the ordinance of the Ministry of Health GM 1,391/05, addressing their opinions and their wishes on the assistance provided to them.

## METHODS

It is a cross-sectional, descriptive study with quantitative-qualitative approach. It had by scene the Hospital of the Foundation of Hematology and Hemotherapy of Pernambuco (Hemope), in Recife, Brazil, with four specialized beds for patients with SCD. Created on November 25, 1977, Hemope Foundation is an organization of scientific, educational and assistance, linked to the State Department of Health (SES). Acting in the areas of Hematology, Hemotherapy, teaching and research, Hemope seeks to meet the health needs of the population in blood transfusion, diagnosis and treatment of blood diseases.<sup>9</sup> By the end of 2014, 773 patients with SCD and age greater than or equal to 18 years old were treated at Hemope, making this service a reference in the treatment of the disease.<sup>10</sup>

The study sample consisted of twenty-six volunteers with hemoglobinopathy and the inclusion criteria were those who were in outpatient care or hospitalized at Hemope Foundation Hospital, aged 18 years or older, living in the State of Pernambuco and accepted to participate in the research by signing the Term of Free and Informed Consent (TCLE). On the other hand, those who were excluded were those who lived outside the state of Pernambuco, who were not assisted by any FHU, or who had any limiting conditions that prevented them from participating in the study.

The action strategy consisted in the analysis of primary data that were collected in the month of August, 2015, through an individual interview conducted by the researcher.

The quantitative variables referred to social and demographic data (age, sex, color or race, marital status, occupation, employment relationship, family income and schooling), variables of housing conditions (type of house, access to basic sanitation, water treatment), lifestyle variables (physical exercise and smoking) and variables on the levels of preventive care offered at FHU (early diagnosis, access to health education, access to medicines, immunobiologicals and home visits). The levels of Health Prevention used were established by Leavell & Clark (1976): primary, secondary and tertiary, divided into five distinct components: promotion, prevention, early diagnosis, treatment and rehabilitation of health problems.

The quantitative analysis was performed using parameters based on relative and absolute measures, and statistical associations of bivariate analysis, as well as graphs and tables constructed with the aid of the Excel 2003 and EPI INFO version 3.5.2 programs with a significance level of 5%, assuming as hypothesis "Regarding the perspective of the SCD carrier, the care provided to him in primary care does not follow the current legislation in the midst of the levels of prevention established by Leavell and Clark."

The qualitative analysis was based on the guiding question "What do you think about the assistance provided by your Health Center?", Where Health Center is referred to the Family Health Unit.

The responses of the participants were transcribed in full for the Word 2007 program and analyzed according to the content analysis of Bardin (2009), which allows a qualitative approach through testimonials as raw material.<sup>11</sup> After saturation of the answers, fifteen lines divided into two thematic categories were selected.

A system of categories is necessary for the codification of content analysis, which categorization has as its primary objective to provide, through condensation, a simplified representation of the collected data. Classification of elements into categories provides the investigation of what each has in common with others, allowing grouping through common parts. In this way, content analysis implicitly determines the belief that categorization does not introduce deviations in the material but that it reveals invisible indices at the raw data level.<sup>11</sup>

The study was carried out respecting Resolution 466/2012 of the National Health Council of the Ministry of Health,<sup>12</sup> and approved by the Research Ethics Committee of Hemope Foundation Hospital under CAAE: 45243015.7.0000.5195. As for the research participants, the secrecy and anonymity were maintained, their names being replaced by black personalities, justified by this disease being more prevalent in this race, and in honor of those who faced the barriers historically built by prejudice.

## RESULTS AND DISCUSSION

Among the 100% (26) of participants who formed the study population, 57.7% (15) belonged to the male sex. About the age group, 46.2% (12) were between 18 and 25 years old, with a mean age of 26 years. In 2010, the life expectancy of these patients in our country was 45 years,<sup>13</sup> which is significantly lower than that estimated for the same individuals living in developed countries (such as the United States), where life expectancy may reach 60 years thanks to the public health policies directed to this group and implemented more than 35 years ago.<sup>1,14</sup>

Regarding color/race, 26.9% (7) declared blacks and 65.4% (17) browns. The Centers for Disease Control and Prevention (CDC) estimates the occurrence of sickle cell disease in 1 in 500 babies born to blacks or African Americans and 1 in 36,000 born among Hispanic Americans.<sup>15</sup> The other sociodemographic data are summarized in Table 1.

**Table 1 -** Sociodemographic data of patients with sickle cell disease participating in the study. Recife, Pernambuco, 2015

Variable	N	%
<b>Age</b>		
18 - 25	12	46,2
26 - 30	06	23,1
31 - 35	07	26,9
36 - 40	01	3,8
<b>Marital status</b>		
Married	6	23,1
Separated	2	7,7
Single	18	69,2
<b>Color/Race</b>		
Black	7	26,9
Brown	17	65,4
White	1	3,8
Indigenous	1	3,8
<b>Education</b>		
Elementary School	7	26,9
High School	17	65,4
College	2	7,7
<b>Family Income*</b>		
Up to one salary	12	46,2
1-3 salaries	14	53,8
<b>Total</b>	<b>26</b>	<b>100</b>

\*Calculation made taking into account the minimum wage of R\$788.00 in the year 2015.

In relation to the occupation, 50% (13) had no employment relationship, receiving only government benefits. 76.9% (20) of the participants lived in the metropolitan region, 96.2% (25) reported living in brick houses and 76.1% (19) had access to basic sanitation.

Of the living habits, 84.6% (22) consumed mineral water, and only 26.9% (7) practiced physical exercises frequently. None of the participants declared themselves smokers. The social determinants of health express the concept that the living conditions are directly related to the health situation of the individual.<sup>16</sup>

Of all participants, only 26.9% (7) were diagnosed by the foot test, and 71.4% (5) were less than 25 years old, as observed in Table 2:

**Table 2 -** Diagnostic means for sickle cell disease to which the study participants were submitted. Recife, Pernambuco, 2015

Age	Foot Test		Other Mean		Doesn't know		Total	
	N	%	N	%	N	%	N	%
18-25	5	41,7	6	50	1	8,3	12	46,2
26-30	2	33,3	4	66,7	0	0	6	23,1
31-35	0	0	7	100	0	0	7	26,9
36-40	0	0	1	100	0	0	1	3,8

These data, through the implementation of ordinance No 822 of June 6, 2001, could be strengthened by the detection of suspected cases, diagnostic confirmation, follow-up and treatment of sickle diseases as recommended by the National Neonatal Screening Program. The Family Health Unit has a fundamental role in this early diagnosis, since it is up to the it to collect or refer clients for the examination.<sup>17</sup>

Early diagnosis is one of the main assumptions of basic care, fitting into the concept of primary prevention, which is nothing more than a set of actions that aim at health promotion, and whose purpose is to reduce or cancel the emergence of comorbidities and pathologies in individuals, alerting them to the risk factors.<sup>7</sup> It is through this early diagnosis that the SCD carriers should be integrated into the SUS care network, thus ensuring the continuity of care as recommended by Ordinance 1.391/05.<sup>6</sup>

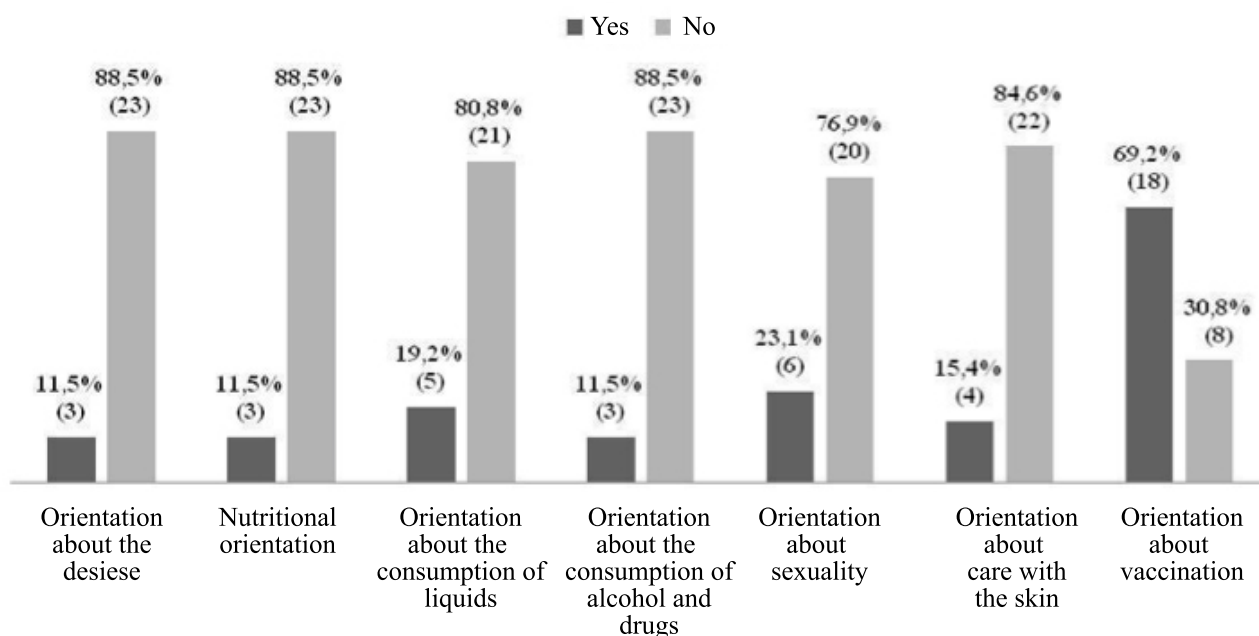
Another important level of primary prevention is health education, which has a set of strategies to promote quality of life, based on the possibility of performing health interventions involving individuals, families and

communities. Such interventions may occur through the articulation of technical and popular knowledge, institutional and community resources, public and private initiatives, encompassing multi-determinants of the health-illness-care process.<sup>18</sup>

In this context, in spite of the knowledge about the SCD, it was investigated through lectures and activities provided in the FHUs, where only 11.5% (3) of the participants reported receiving some kind of information about the disease. In Figure 1, we observed the guidelines regarding factors associated with SCD, to which 11.5% (3) reported having received nutritional guidelines and 65.4% (17) had their body mass index (weight-height) measured in the Unity as a complementary action.

This situation is aggravated, since 50% (13) of the interviewees claimed that they are not the only people living with SCD living in their community, which demonstrates once again the importance of educational actions aimed at the health of these patients.

**Figure 1** - Health education actions dedicated to the study participants in the FHU who are assisted. Recife, Pernambuco, 2015





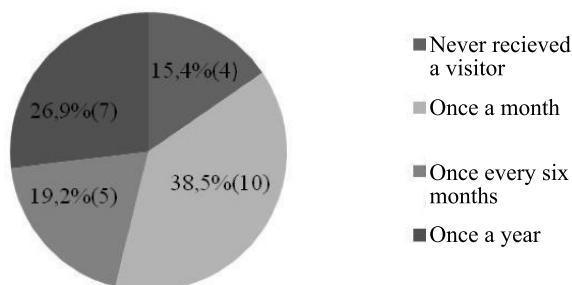
Health education is recommended by the decree 1,391/05 through paragraph II, which guarantees the promotion of access to information to the SCD, aiming to strengthen this target public.<sup>6</sup> In addition, one of the main goals of health education is to make the student a more autonomous being, allowing him to make choices that will change his quality of life. To increase this potential, what the learner needs is effective practices on the part of health professionals, supporting them in their own care.<sup>19</sup>

The accomplishment of educational activities provides a greater approximation of the multiprofessional health team to the person with SCD, demonstrating that this is a good motivational tool. It is important that educational practice be carried out continuously, regardless of the economic and human resources available, in order to form bonds of trust with the clients.<sup>20</sup>

In addition, in performing their role as educator, regardless of the level of performance or modality of treatment, the nurse contributes so that the person living with sickle cell disease has a less traumatic life and has a smoother prognosis.<sup>21</sup>

Regarding the home visit performed by the Community Health Agents (CHAs), the results can be observed in Figure 2:

**Figure 2** - Frequency of home visits received by research participants. Recife, Pernambuco, 2015



According to Ordinance 2.488/11, the home visit is one of the attributes of the CHA and should be performed at least once a month according to the need of each individual. This practice is considered a good resource for the knowledge of families, providing observing the individual in their environment. The repetition of visits makes routine contact creating links, facilitating the identification of conditions that can lead to illness.<sup>22,23</sup>

Regarding the counter-reference, none of the participants were referred for ophthalmologic examination through FHU, only 3.8% (1) stated that they had already undergone radiological examinations and 11.5% (3) to perform ultrasonography, all of them being women.

One of the most important points of the FHS is to be a guiding force in the health care network, as well as being resolute in the referral and counter-referral system, since among other factors, it facilitates the flow of referral of users

to the different levels of attention. The non-structuring of this system in the health services makes it impossible to continue health care, preventing the assistance to follow its flow in the other levels of complexity.<sup>22,24</sup>

Access to health services is related to factors according to the needs of demand and supply. This concept of access can be broadened to contextual factors such as health policies and the provision of services.<sup>25</sup> With this concept in mind, 61.5% (16) of the interviewees reported that they attend FHU.

Below are the categories referring to “(de)construction of the link related to the knowledge of professionals”, which express well the reflection of primary care in the Family Health Unit that provides care.

*“I like my Center. They do not know much about sickle cell anemia, but they always treat me well.” (Nelson Mandela)*

*“My Center is great. It offers me full support and thanks to that I do not need to go so much to the Hemope.” (Luther King)*

*“I know what I have and I know that the Center will not know how to take care of me.” (Joaquim Barbosa)*

*“I do not go because they do not know my disease! The Health Agent from my Center spread to the people on my street that sickle cell anemia was contagious, so the kids did not play with me.” (Bob Marley)*

Traditionally, the treatment and care of patients with sickle cell disease are delegated only to haematological centers. Because of this, intermediate levels of health care are unaware or even ignorant of the disease. This perception has been altered through the premise that sickle cell disease is a chronic pathology that affects all spheres of the patient's life, that is, not only their physical condition, but also their social, family and work interactions. Although there are health policies for people with sickle cell disease, these alone are not enough, requiring the commitment of health professionals to receive these users.<sup>25,26</sup>

*“They do not know anything about what I have. I had to bring Hemope pamphlets so they would understand.” (Jesse Owens)*

*“I like my nurse. The people know what I have and take care of me.” (Machado de Assis)*

*“[...] I do not like it and I do not trust the people there. I'm only going because I have to do the dressing.” (Rosa Parks)*

*"I like my Center. The Health Agents treat me well and the nurse is great. She knows what I have and so she knows how to take better care of me."* (Muhammad Ali)

The Family Health Strategy (FHS) should offer its clients direct assistance through the control of the health-disease process and also in the management. The FHS team must be formed by at least one doctor, one nurse, one auxiliary or nursing technician and Community Health Agents (CHA). To this composition, the oral health professionals (or Buccal-eSB team: dental surgeon, auxiliary and/or technician in Oral Health) may be added to this composition.<sup>27,28</sup>

Faced with this scenario, questioned about the care by multiprofessional teams, only 34.6% (9) of the participants stated that they were accompanied by other professionals who were not doctors or nurses.

It is also possible to observe in the participants' discourses one of the main reasons for attending or not the FHU, ranging from the lack of adequate infrastructure to the professionals' knowledge about sickle cell disease care, as established by Decree 1391/05.

From this premise are the categories related to accessibility in primary health care:

*"I'm just going to pick up materials from my dressing."* (Jesse Owens)

*"[...] But sometimes material is lacking."* (Machado de Assis)

*"The nurse at my Center said that folic acid was only for pregnant women, and that I should buy it."* (Dandara)

*"Terrible employees. A nurse thought I was pregnant because I was always going to get folic acid and asked if my son was never going to be born."* (Michelle Obama)

*"The people there do not treat me very well. I'm only going to get medicine, but it's usually lacking."* (Ernesto Carneiro Ribeiro)

The reception of SCD carriers by the FHS is guaranteed by Ordinance 1,391/05, which advocates for a comprehensive care through the services provided by a multidisciplinary team.<sup>6</sup>

The decree 1,391 guarantees to the patients of SCD access to the essential medicines, according to protocol of special immunobiological and inputs.<sup>6</sup> In addition, it is the Primary Care's responsibility to offer basic and special immunizations, as well as the prescription and dispensing of medications, such as: folic acid, penicillin or other antibiotics, analgesics and anti-inflammatories. It is

expected that 80% of people with SCD are followed up in primary health care.<sup>29</sup> However, in the current study, 69.2% (18) reported having access to medication in the FHU, but are treated in a discriminated manner.

At the end of the day, the participants were asked to give a score of 0 to 10 for the FHU which served them. The average number of grades was 3.7, showing that the service has a risk of stigmatizing the perception triggered by a discrediting of the quality of public health establishments.

## CONCLUSION

The research findings reflect the dissatisfaction and low credibility that the majority of patients of SCD have in relation to primary care. It would be desirable for the ESF to strengthen ties with the attached population, enabling assistance and strengthening the bond with the service.

There is a need for improvement in the care and reception of these clients, since sickle cell disease is considered a public health problem in our country, respecting three principles that underlie the Unified Health System: universality, completeness and equity.

It is desirable that there is a greater interaction of these patients with the community, and this makes the FHU a gateway to social interaction, making it possible to share experiences through the creation of groups. The establishment of a new relationship between health professionals and the community is of an urgent nature, in an attempt to change this image that sickle cell patients have about the service.

Therefore, it is concluded that the premises established by Ordinance GM 1,391/05 are heading to be achieved, even after ten years of its implementation. Public policies aimed at the black population need to be rescued by the FHS, so that there is continuity of care.

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